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The abdominal cocoon—an unusual cause of intestinal obstruction in adolescents

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Summary The abdominal cocoon is a rare cause of intestinal obstruction in adolescent girls caused by encasement of the small intestine in fibrous tissue. Only 15 cases have been reported in the English literature so far. This is a report of another two patients managed at the Aga Khan University Hospital. The correct diagnosis is often not suspected, resulting in delays in the treatment of this condition. Following simple surgical release of the entrapped bowel, these patients do well.

Introduction

The abdominal cocoon is a condition in which a variable length of healthy small bowel is enveloped in a fibrocollagenous membrane, giving the appearance of a cocoon. It is an unusual cause of intestinal obstruction in children and adolescents, and has received little attention in standard paediatric surgical literature.[1,2] Until February 1991, 15 children with an abdominal cocoon had been reported in the English literature.[3] We report two additional cases who were managed recently at the Aga Khan University Hospital.

Case No. 1

A 13-year old girl was admitted for evaluation of abdominal pain, vomiting and constipation of 48 hours duration. She had a history of intermittent abdominal discomfort for 1 month, for which she had received spasmolytics from general practitioners. She had had normal menstrual periods for 1 year.

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Physical examination revealed a mildly dehydrated patient with a distended and diffusely tender abdomen with no palpable mass. On rectal examination there was no abnormality. The laboratory findings included a haemoglobin level of 12 g/dl, leucocyte count of 12,800/mm^3 and an erythyrocyte sedimentation rate (Westergren) of 10 mm/h. Blood glucose, serum electrolytes and liver function tests were within normal limits. An abdominal radiograph revealed dilated loops of small bowel with an obstructive pattern. A diagnosis of mechanical small bowel obstruction was made. Following resuscitation, she underwent an exploratory laparotomy. The jejunum was found to be dilated and the entire ileum discoloured, oedematous and covered by a dense whitish membrane giving the appearance of a cocoon (Fig. 1) and extending over the liver, gall bladder and upper abdominal viscera. The membrane envelopping the small bowel was incized carefully and the coils of intestine separated from each other with the intestinal serosa intact (Fig. 2). The postoperative period was uneventful and the girl was discharged on the 10th day. Microscopic examination of the membrane revealed a

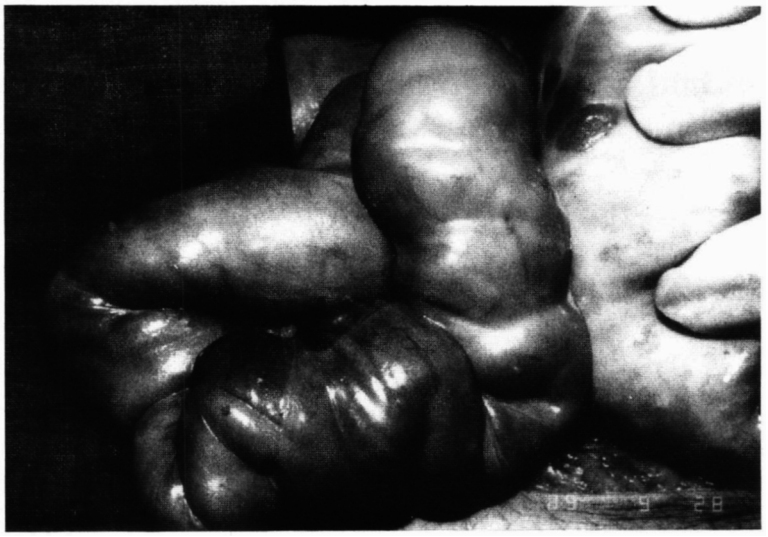

FIG. 1. Operative photograph demonstrating a thick membrane encasing the small bowel.

chronic non-specific inflammatory reaction and absence of mesothelium. A biopsied mesenteric lymph node revealed non-specific sinus hyperplasia and routine and acid-fast bacillus (AFB) culture of the peritoneal fluid was negative. The girl has remained well over 18 months of follow-up.

Case No. 2

A 13-year-old girl presented with a 4-month history of nausea, vomiting, abdominal pain and weight loss. She had been having normal periods for 5 months. On examination, she looked ill and was emaciated. The abdomen was diffusely tender, with a vague mass in the left lower quadrant. An ultrasound examination of the abdomen revealed a localized collection of fluid in the pelvis which, on aspiration and microscopy, showed predominantly lymphocytes and had a glucose level of 37 mg/dl and a protein level of 4000 mg/dl. At this point, the girl left hospital against medical advice. Owing to the persistence of abdominal pain and vomiting despite various medications, she returned to the Aga Khan University Hospital 1 month later and was admitted. Abdominal examination again revealed a mass in the left lower quadrant, and abdominal radiographs showed air fluid levels, suggestive of partial intestinal obstruction. A contrast-enhanced CAT scan of the abdomen showed matted loops of small intestine in the mid- and left-lower abdomen, with free fluid in the pelvis (Fig. 3). At laparotomy, the peritoneal cavity contained a small amount of serosanguinous fluid. The small bowel from the duodenojejunal flexure to the caecum was found to be encased in a thick white fibrous membrane extending onto the upper abdominal and pelvic viscera. Incision of the membrane released the coils of dilated, fluid-filled small bowel. The histology of the excized membrane revealed fibroblastic reaction, chronic non-specific inflammation and the absence of a mesothelial lining. Routine and AFB cultures of the peritoneal fluid were sterile. The girl made an unventful recovery and was discharged on the 5th post-operative day. She has remained well during the 6 months following surgery.

Discussion

The abdominal cocoon is a rare cause of small bowel obstruction in children and adolescents.[4,5] The condition was first described by Foo *et al.*[6] in 1977 and since then a total of 15

FIG. 2. After excision of the cocoon, the loops of small bowel are free to expand

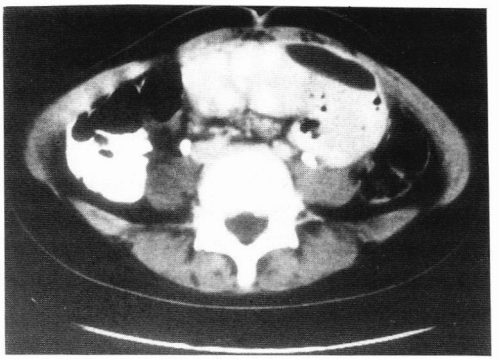

FIG 3. CT scan showing matted loops of small intestine and free fluid in the peritoneal cavity.

cases have been reported in the English literature.[3,7-10] All reports, including the present one, indicate a prevalence of this entity in girls between 4 and 18 years of age, most commonly in those living in tropical and subtropical regions. The condition is characterized by encasement of the small intestine in a concertina-like fashion in a fibrocollagenous membrane with the appearance of a cocoon.[5,6] The white membrane can be incized relatively easily and the bowel loops usually expand in a normal fashion. There appears to be no association of the condition with previous surgery or infections of the peritoneal cavity, and the occasional adhesions between the bowel loops and the peritoneum are easily divided.

This condition must be differentiated from encapsulating sclerosing peritonitis which occurs in association with the use of the beta blocker practolol, cirrhosis of the liver, chronic peritoneal dialysis, carcinoid syndrome, familial Mediterranean fever and asbestos exposure.[11-13] In this condition the entire peritoneal cavity is often obliterated by dense adhesions, making it difficult to enter the abdominal cavity. The visceral and parietal peritoneum are thickened with a shortened small bowel encased in a rigid tube. There is usually no plane of cleavage between the membrane and the bowel, making release of the bowel loops extremely difficult.

The abdominal cocoon is occasionally confused with peritoneal encapsulation, which is an embryological abnormality and usually an incidental finding at laparotomy.[1,9,14] The clinical presentation, however, is markedly different, with eight of the nine cases reported being men in the 6th decade of life. Intestinal obstruction occurred in only one case,[14] unlike in patients with the abdominal cocoon, where the presentation is almost always one of acute or chronic intestinal obstruction in an adolescent girl. At laparotomy, a characteristic sac is

found attached to the colon and the posterior parietal peritoneum and containing a portion of or the entire small bowel from the duodenojejunal junction to the ileocaecal region. Once the sac is removed the bowel is found to be entirely normal, with a normal serosal surface.

The aetiology of the cocoon remains obscure. Absence of a mesothelium in the membrane suggests that it may be the result of progressive connective tissue formation following attenuation of mesothelial cells. The age, sex and geographical distribution of the patients suggest that the abdominal cocoon may be a sequela to subclinical infection of the peritoneum.[3,6,9] The responsible organisms may be endemic with a predilection for the genital tract, leading to retrograde peritonitis via the Fallopian tubes.[3,9] This may explain the occurrence of this condition in females only. The role of retrograde menstruation with superimposed viral infection has been suggested, but the presence of premenarchal patients in some reports appears to refute this hypothesis.

The clinical features of the abdominal cocoon may include anorexia, nausea, vomiting, weight loss, abdominal distension and a palpable abdominal mass. The most common presentation appears to be acute or subacute intestinal obstruction.[3,6,9] A pre-operative diagnosis is almost never made and the non-specific and intermittent symptoms may result in delay in diagnosis, as demonstrated by the second girl in our report. Laboratory and radiological studies are non-specific, although plain radiographs of the abdomen may suggest features of intestinal obstruction. A characteristic serpentine configuration of dilated small bowel within the cocoon may be noted on a barium meal follow-through examination.[9] The second girl in this report underwent a CAT scan of the abdomen, which showed matted small bowel in the mid- and left-lower abdomen, and free fluid in the pelvic cavity. This appearance may be helpful in the diagnosis of the condition.

The treatment of choice is simple incision of the membrane, which frees the dilated small bowel loops.[3,9] Additional procedures, including bowel resection, enteroenterostomy, Noble's plication of the mesentery and insertion of long intestinal tubes have been reported,[2,5–7,9] but may increase morbidity and are probably unnecessary. An incidental appendectomy is recommended, as the appendix would be difficult to find should the patient develop acute appendicitis later on. The long-term prognosis is excellent, with death reported in only one patient who had long-standing symptoms and weight loss.[6]

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